

NOTES

PULMONARY VASCULAR DISEASE

GENERALLY, WHAT IS IT?

PATHOLOGY & CAUSES

- Diseases affecting blood flow through pulmonary vasculature, or fluid flow from vasculature
- Can be caused by process within lungs/ elsewhere in body

SIGNS & SYMPTOMS

- Dyspnea, poor effort tolerance, chest pain, tachypnea

DIAGNOSIS

X-ray, chest CT scan, spirometry, ultrasound, echocardiogram, ECG

TREATMENT

- Supportive, treat underlying disease, optimize organ function (heart, lungs)

CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION

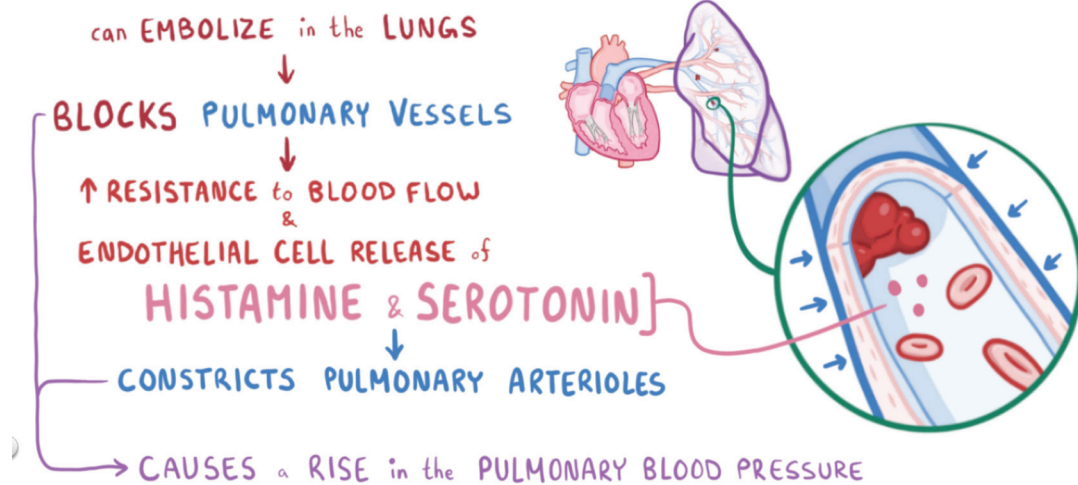


Figure 129.1 Chronic thromboembolic pulmonary hypertension is an example of a pulmonary vascular disease that originates outside the lungs. In this case, an embolism blocks the pulmonary vessels, causing pulmonary blood pressure to rise beyond normal levels.

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PULMONARY EDEMA

osms.it/pulmonary-edema

PATHOLOGY & CAUSES

- Alteration in Starling forces → build up of fluid within interstitial space, air spaces of lung

CAUSES

Cardiogenic (heart disease)

- Left sided heart failure → inefficient pumping of blood from heart by left ventricle → blood backs up into left atrium → pulmonary circulation → pulmonary hypertension (raised hydrostatic pressure) → more fluid in lung interstitium → pulmonary edema
 - Severe systemic hypertension (> 180/110mmHg) → left ventricle cannot pump effectively against extreme afterload → blood backs up into left atrium → pulmonary circulation → pulmonary edema

Non-cardiogenic (damage to pulmonary capillaries or alveoli)

- Direct damage to alveoli/vasculature → inflammatory response → leaky capillaries
 - Pulmonary infection, toxin inhalation, chest trauma, pulmonary vein occlusion, burns
 - Sepsis → systemic inflammation → global edema
 - Insufficient circulation of osmotically active proteins, e.g. albumin → low oncotic pressure in capillaries
- Malnutrition
- Liver failure
- Excessive protein loss (nephrotic syndrome, protein losing enteropathies)

COMPLICATIONS

- **Impaired gas exchange:** oxygen/carbon dioxide must diffuse through wide layer of fluid → blood unable to fully saturate

- Free fluid predisposes to secondary infection

SIGNS & SYMPTOMS

- Dyspnea, productive cough (pink frothy sputum), excessive sweating, anxiety, tachycardia, end-inspiratory crackles, dullness to percussion, cyanosis (decreased hemoglobin saturation)
- Pulmonary edema in heart failure may also include
 - Orthopnea (shortness of breath worse when lying flat)
 - Paroxysmal nocturnal dyspnea (episodes of severe sudden breathlessness at night)
 - Peripheral pitting edema
 - Raised jugular venous pressure
 - Hepatomegaly

DIAGNOSIS

DIAGNOSTIC IMAGING

Chest X-ray

- Kerley B lines (thickened subpleural interlobular septa, usually seen at base of lung)
- Increased vascular shadowing → batwing perihilar pattern
- Upper lobe diversion (prominent upper lobe pulmonary veins)
- Pleural effusion (if edema severe)

Non-contrast high resolution chest CT scan

- Airspace opacity
- Smooth thickening of interlobular septae

Chest ultrasound

- Detection of small amounts of fluid
- Echo-free space between visceral and parietal pleura
- Septations in pleural fluid → underlying

infection, chylothorax/hemothorax

Echocardiograph

- Evaluation of cardiac function, can demonstrate left ventricular failure

LAB RESULTS

- Serum electrolytes
- Renal function
- Inflammatory markers
- Low oxygen saturation
- Increased carbon dioxide

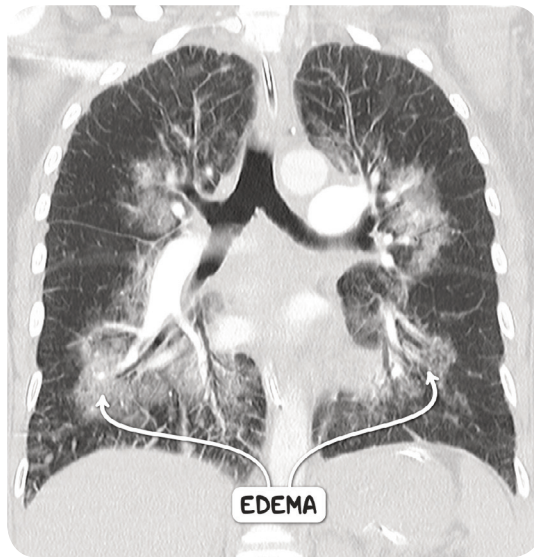


Figure 129.2 A CT scan of the chest in the coronal plane demonstrating the peribronchovascular distribution of acute pulmonary edema.

PULMONARY EDEMA

↳ EXCESS FLUID in the PULMONARY INTERSTITIUM
↳ POOR GAS EXCHANGE

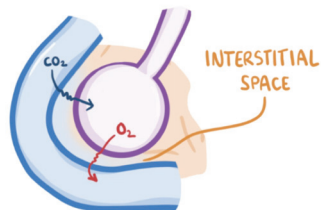


Figure 129.4 Illustration depicting pulmonary edema.

TREATMENT

MEDICATIONS

- If cardiogenic
 - *Preload reduction*: nitroglycerin, diuretics, morphine sulphate
 - *Afterload reduction*: ACE inhibitors, angiotensin II receptor blockers, nitroprusside
- If non-cardiogenic
 - Manage illness (e.g. treat infection)

OTHER INTERVENTIONS

- Continuous positive airway pressure (CPAP)
- *Intubation*: mechanical ventilation if level of consciousness compromised

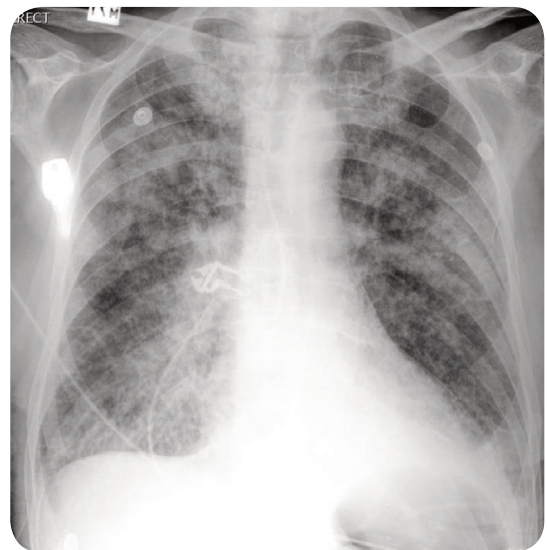


Figure 129.3 A plain chest radiograph demonstrating pulmonary edema. There is interstitial edema, represented by fine stranded opacities known as Kerley B lines, as well as alveolar edema, represented by confluent nodular opacities.

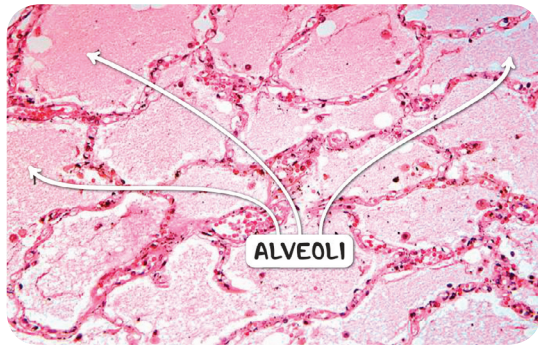


Figure 129.5 The histological appearance of pulmonary edema.

PULMONARY EMBOLISM

osms.it/pulmonary-embolism

PATHOLOGY & CAUSES

- Blockage of pulmonary artery by a substance brought there via bloodstream
- Thrombus in remote site **embolizes** → lodges in pulmonary vascular tree → “pulmonary embolism”
- Obstruction of blood flow distal to embolism → increased pulmonary vascular resistance → increased pulmonary artery pressure → increased right ventricular pressure → cor pulmonale (if severe obstruction)
- **Regional decrease in lung perfusion** → dead space (ventilation, but no perfusion) → hypoxemia → tachypnea

Source of embolus

- **Lower extremity deep vein thrombosis**
 - Most arise from deep veins above knee, iliofemoral deep vein thrombosis
 - Can arise from pelvic deep veins
 - Pelvic thrombi tend to advance to more proximal veins before embolizing
- Upper extremity deep veins (rarely)
- **Uncommon embolic material:** air, fat, amniotic fluid

RISK FACTORS

- **Virchow’s triad:** endothelial injury, stasis of blood flow, blood hypercoagulability
- > 60 years old, malignancy, history of deep vein thrombosis/pulmonary embolism, hypercoagulable states, genetic disorders (e.g. Factor V Leiden thrombophilia), dehydration, prolonged immobilization (bed rest, travel), cardiac disease, obesity, nephrotic syndrome, major surgery, trauma, pregnancy, estrogen-based medication (e.g. oral contraceptives)
- Increased risk of fat embolism with bone fractures (e.g. hip, femur)

SIGNS & SYMPTOMS

- **Dyspnea, pleuritic chest pain, cough, hemoptysis**
- Signs, symptoms of deep vein thrombosis
 - Tender, swollen, erythematous extremity
- Syncope
- Often asymptomatic (in the case of small emboli)



MNEMONIC: TOM SCHREPFER

Risk factors for Pulmonary embolism

Trauma
Obesity
Malignancy
Surgery
Cardiac disease
Hospitalization
Rest (bed-ridden)
Elderly
Past history
Fracture
Estrogen (pregnancy, post-partum)
Road trip

- Low SpO₂, tachypnea, rales, tachycardia, S4 heart sound, increased P2 (closure of pulmonary valve), shock, low-grade fever, decreased breath sounds, percussion dullness, pleural friction rub, sudden death (pulmonary saddle embolism)

DIAGNOSIS

Wells' score

- Used to assess probability of pulmonary embolism (multiple different probability tests available)
 - Score > 4: pulmonary embolism likely, consider diagnostic imaging
 - Score ≤ 4: pulmonary embolism unlikely, consider D-dimer test to rule out

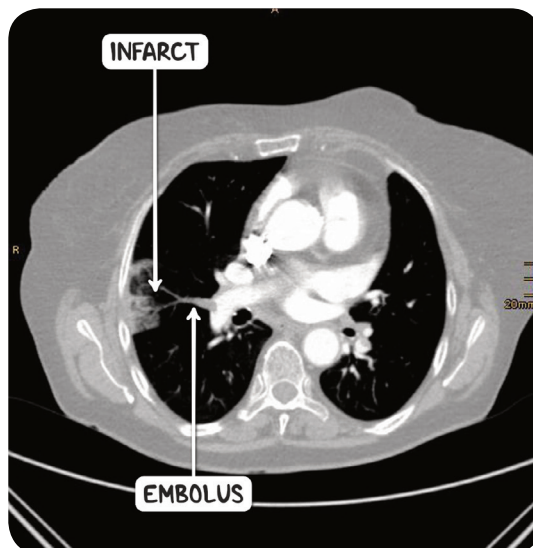


Figure 129.6 A CT pulmonary angiogram demonstrating a pulmonary embolus and subsequent right middle lobe infarct.

WELLS' SCORE

CRITERIA	POINTS
CLINICAL SUSPICION OF DEEP THROMBOSIS	+3
ALTERNATIVE DIAGNOSIS LESS LIKELY THAN PULMONARY EMBOLISM	+3
TACHYCARDIA PRESENT	+1.5
IMMOBILIZATION (≥ 3 DAYS) & SURGERY IN PAST 4 WEEKS	+1.5
PRIOR HISTORY OF DEEP VEIN THROMBOSIS OR PULMONARY EMBOLISM	+1.5
HEMOPTYSIS	+1
MALIGNANCY (OR ≤ 6 MONTHS OF TREATMENT)	+1

DIAGNOSTIC IMAGING

Chest X-ray

- Typically normal

CT pulmonary angiography

- Definitive test
- Visualize decreased blood supply

Venous duplex ultrasound

- Of lower extremities
 - May reveal origin of pulmonary embolism
 - Negative result does not exclude pulmonary embolism

Ventilation-perfusion scan

- Normal scan rules out pulmonary embolism

LAB RESULTS

- D-dimer** (high negative predictive value)
 - Positive result does not prove pulmonary embolism
 - Negative result rules out pulmonary embolism
- Arterial blood gas**
 - $\downarrow$ PaO₂ → hypoxemia
 - Hyperventilation → $\uparrow$ PaCO₂ → $\uparrow$ pH → respiratory alkalosis
 - A-a gradient elevated (indicated V/Q mismatch)
- Tests for causes of secondary pulmonary embolism
 - Full blood count, clotting profile, erythrocyte sedimentation rate, renal function, liver function, electrolytes

OTHER DIAGNOSTICS

ECG

- Excludes other causes of chest pain
- ECG features of pulmonary embolism (or any pulmonary hypertension) include
 - Sinus tachycardia
 - Right bundle branch block
 - Right ventricular strain pattern:** T wave inversion in right precordial (V₁–V₄), and inferior leads (II, III, aVF)
 - Right atrial enlargement (P pulmonale)
 - Right atrial dilatation → right axis deviation

- Dominant R wave in V₁
- S₁Q₃T₃ pattern:** Deep S wave in lead I, Q wave in lead III, negative wave in lead III
- Nonspecific ST segment, T wave changes
- Pulmonary embolism can be excluded if
 - SaO₂ exceeds 95%
 - Age < 50
 - No unilateral leg swelling, hemoptysis, history of deep vein thrombosis/pulmonary embolism, recent surgery/trauma, hormone use (or estrogen-based medications), tachycardia

TREATMENT

MEDICATIONS

Anticoagulation

- Acute phase (days–weeks)
 - Prevent further thromboembolic events
 - Unfractionated heparin, low molecular weight heparin, fondaparinux
- Long-term (vitamin K antagonists)
 - Warfarin, acenocoumarol, phenprocoumon

Thrombolysis

- Used for massive pulmonary embolism causing hemodynamic instability
- Carries risk of secondary hemorrhage
- Thrombolytics used to break up clots
 - Streptokinase, staphylokinase, urokinase, anistreplase
 - Recombinant tissue plasminogen activators (alteplase, reteplase, tenecteplase)

SURGERY

Pulmonary thromboendarterectomy

- Surgical removal of a chronic thromboembolism
- Rare

Inferior vena cava filter

- Vascular filter inserted into inferior vena cava to prevent life-threatening pulmonary emboli
- Indications: anticoagulant therapy contraindicated, major embolic event

despite anticoagulation

OTHER INTERVENTIONS

Preventative measures

- Unfractionated heparin, low molecular weight heparin
- Factor Xa inhibitor
- Long-term low-dose aspirin
- Anti-thrombosis compression stockings/ intermittent pneumatic compression

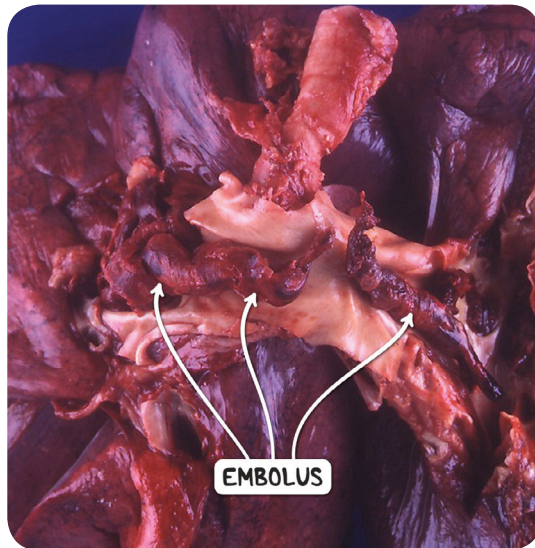


Figure 129.8 The gross pathological appearance of a pulmonary embolus.

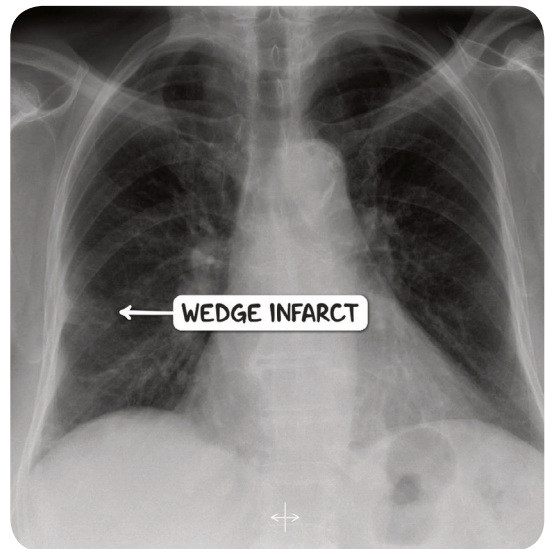


Figure 129.7 A plant chest radiograph of the same individual, demonstrating the pulmonary infarct which is visible as a wedge shaped opacity in the lateral art of the right lung field.

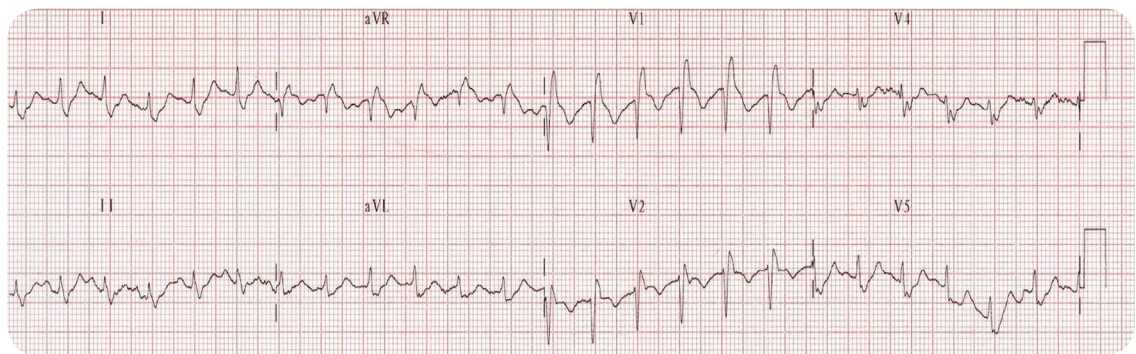


Figure 129.9 The ECG changes associated with a pulmonary embolism. There is a right bundle branch block, sinus tachycardia and T-wave inversions in leads V1-3 and III.

PULMONARY HYPERTENSION

osms.it/pulmonary-hypertension

PATHOLOGY & CAUSES

- Increased blood pressure in pulmonary circulation
- Mean pulmonary arterial pressure > 25mmHg (normal ~15mmHg)
- Pulmonary hypertension → excess fluid in pulmonary interstitium (pulmonary edema) → impaired gas exchange
- Pulmonary hypertension → strain on right heart → hypertrophy → right heart oxygen demand eventually exceeds supply → right-sided heart failure
 - Right heart failure caused by lung disease → cor pulmonale → backup of blood in venous system → signs, symptoms of right heart failure
- Raised jugular venous pressure
- Fluid build up in liver → hepatomegaly
- Fluid build up in legs → leg edema
- Left ventricle receives less blood → compensation → pumps harder, faster (tachycardia)
- Idiopathic, inherited, drug/toxin associated causes connective tissue disease, HIV infection, portal hypertension congenital heart disease (shunting)

Group II

- Pulmonary hypertension secondary to left heart disease
- Pulmonary hypertension due to left heart disease (heart failure, valvular dysfunction) → left heart fails to pump blood efficiently → backup of blood in pulmonary veins, capillary beds → increased pressure in pulmonary artery → pulmonary edema, pleural effusion
- Raised back pressure may trigger secondary vasoconstriction → increased right heart strain
- Common causes include
 - Left ventricular systolic/diastolic dysfunction
 - Valvular heart disease
 - Congenital/acquired in/out-flow tract obstruction
 - Congenital cardiomyopathy
 - Pulmonary venous stenosis

TYPES

Group I

- Pulmonary arterial hypertension, pulmonary veno-occlusive disease, pulmonary capillary hemangiomatosis
- Abnormal increase in pulmonary arteriolar resistance → increased strain on right heart (pumping fluid through narrower pipe)
- Damage to endothelial cells lining pulmonary arteries → release of endothelin-1 serotonin, thromboxane, produce less nitric oxide and prostacyclin → constriction of arterioles, hypertrophy of smooth muscle → pulmonary hypertension
- Over time affected vessels become stiffer, thicker (fibrosed) due to vasoconstriction, thrombosis, vascular remodeling → greater increase in blood pressure in lungs, more strain on right heart

Group III

- Pulmonary hypertension due to lung disease/chronic hypoxia
- Low oxygen levels in alveoli pulmonary arteries constrict
- Chronic lung disease → region of diseased lung → inefficient/total lack of gas exchange → hypoxic vasoconstriction (pulmonary arterioles) → shunting of blood away from damaged areas
- Prolonged alveolar hypoxia across wide portion of pulmonary vascular bed → increase in pulmonary arterial pressure → thickening of pulmonary vessel walls → greater effort required from right heart → sustained pulmonary hypertension
- Causes include
 - COPD

- Interstitial lung disease
- Mixed restrictive/obstructive pattern disease
- Sleep-disordered breathing
- Alveolar hypoventilation
- Chronic exposure to high altitude

Group IV

- **Chronic arterial obstruction/thromboembolic disease**
- Recurrent blood clots in pulmonary vasculature
- Blockage/narrowing of pulmonary vessel with unresolved obstruction (e.g. clot) → increased pressure, shear stress (turbulence) in pulmonary circulation → vessel wall remodelling → sustained pulmonary hypertension
- Causes endothelium to release histamine, serotonin → constriction of pulmonary arterioles → rise in pulmonary blood pressure → chronic thromboembolic pulmonary hypertension
- Other causes of arterial obstruction
 - Angiosarcoma, arteritis, congenital pulmonary artery stenosis, parasitic infection

Group V

- **Unclear/multifactor mechanisms**
- Hematologic disease (e.g. hemolytic anemia)
- Systemic disease (e.g. sarcoidosis, vasculitis)
- Metabolic disorders (e.g. glycogen storage disease, thyroid disease)
- Other (e.g. microangiopathy, chronic kidney disease)

RISK FACTORS

- Family history, prior pulmonary embolic events, HIV/AIDS, sickle cells disease, cocaine use, COPD, sleep apnea, living at high altitude, mitral valve pathology

SIGNS & SYMPTOMS

- Dyspnea, syncope, fatigue, chest pain, poor effort tolerance, loss of appetite, lightheadedness, orthopnea (left-sided heart failure)
- Tachycardia, cyanosis, parasternal heave
- **Signs of systemic congestion/right heart failure:**
 - Loud pulmonic component of second heart sound (P_2)
 - Jugular venous distension
 - Ascites
 - Hepatojugular reflux
 - Lower limb edema

DIAGNOSIS

DIAGNOSTIC IMAGING

Chest X-ray

- **Enlarged pulmonary arteries**
- Lung fields may or may not be clear, dependent on underlying cause

Echocardiogram

- **Increased pressure** in pulmonary arteries, right ventricles → **dilated pulmonary artery**
- **Dilatation/hypertrophy** of right atrium, right ventricle
- **Large right ventricle** → bulging septum

Ventilation/perfusion scan

- Identify / exclude ventilation-perfusion mismatches

OTHER DIAGNOSTICS

Right heart catheterisation (gold standard)

- Catheter into right heart → most accurate measure of pressures

ECG

- **Right heart strain pattern:** T wave inversion in right precordial (V_1 – V_4), and inferior leads (II, III, aVF)

Spirometry

- Unidentified underlying cause

TREATMENT

MEDICATIONS

- Pulmonary hypertension secondary to left ventricular failure → optimize left ventricular function
 - Diuretics (cautiously—individuals may be preload dependent)
 - Digoxin
 - Anticoagulants
- Cardiogenic pulmonary arterial hypertension
 - Relax smooth muscle (promote vasodilation), reduce vascular remodelling, improve exercise capacity

with prostanoids, phosphodiesterase inhibitors, endothelin antagonists

- Pulmonary arterial hypertension
 - Endothelin receptor antagonists
 - Prostanoids

SURGERY

- Lung transplant
- Repair/replace damaged valves to optimize left ventricular function

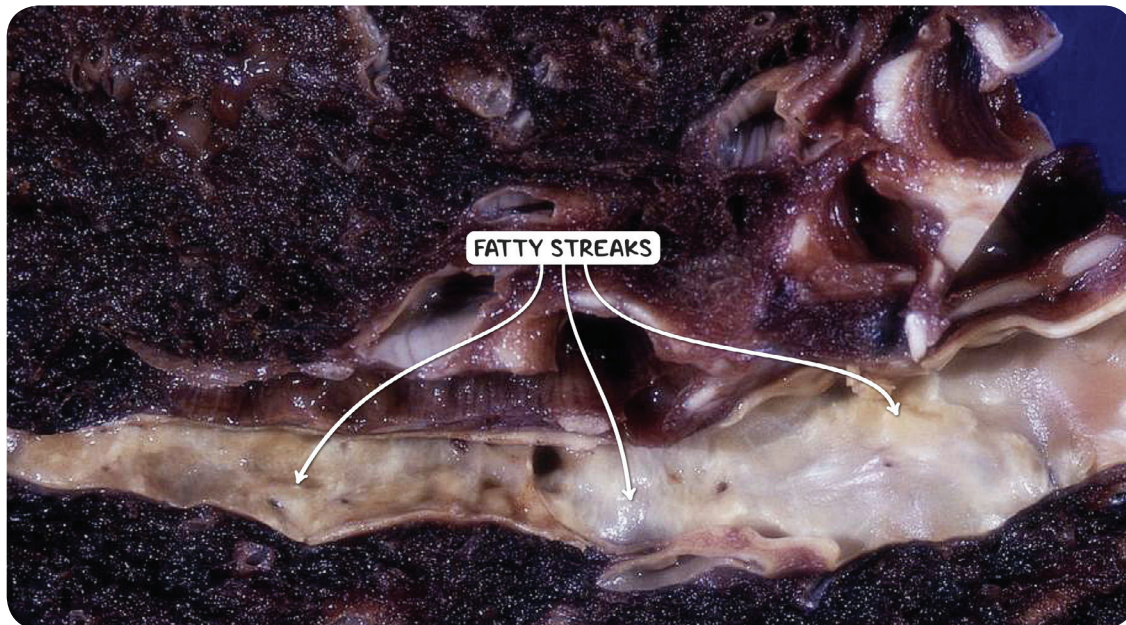


Figure 129.10 The gross pathological appearance of the pulmonary arteries in a case of pulmonary hypertension. The underlying pathological process is similar to atherosclerosis found elsewhere in the cardiovascular system.

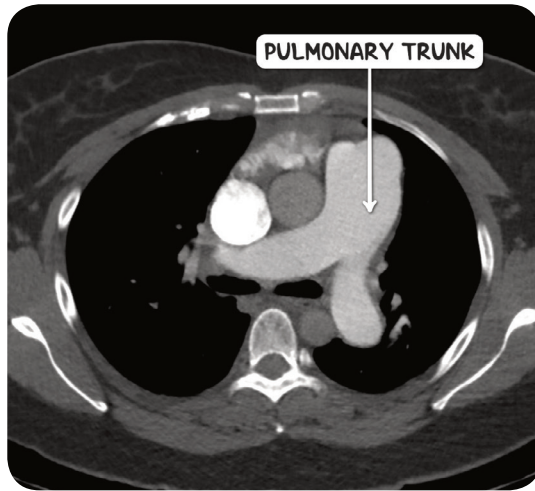


Figure 129.11 A CT scan of the chest in the axial plane demonstrating enlargement of the pulmonary trunk as a consequence of pulmonary hypertension.

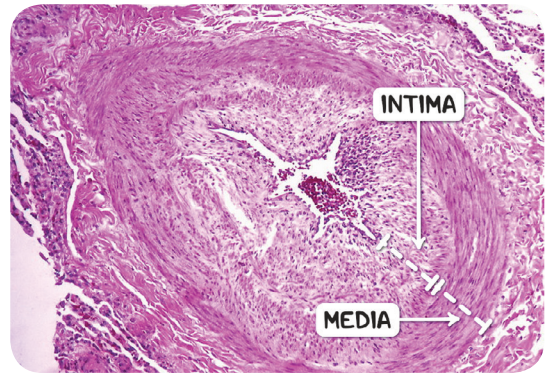


Figure 129.12 The histological appearance of a pulmonary artery in a case of pulmonary hypertension. There is marked thickening of both the intima and the media.